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CASE REPORT

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Ischemic stroke in children in course of moyamoya disease – case report

Jerzy Pietruszewski[1], Ewa Jamroz[1], Katarzyna Gruszczyńska[2], Jan Baron[3], Ewa Głuszkiewicz[1], Joanna Gibińska[3]

[1] Clinic of the Developmental Age Pediatrics and Neurology, Medical University of Silesia, Katowice, Poland
[2] Imaging Diagnostics Department, Radiology and Nuclear Medicine Department, Medical University of Silesia, Katowice, Poland
[3] Department of Radiology and Nuclear Medicine, Medical University of Silesia, Katowice, Poland

Author's address: Katarzyna Gruszczyńska, Imaging Diagnostics Department, Radiology and Nuclear Medicine Department, Medical University of Silesia, Medyków 16, 40-752 Katowice, e-mail: kgruszczynska@poczta.onet.pl

Summary

Background: Progressive stenosis of the supraclinoid segments of the internal carotid arteries, followed by formation of characteristic collateral brain circulation is typical for moyamoya disease. This illness, with unknown pathogenesis, is often diagnosed in Asiatic population. In Poland it can be a rare cause of ischemic infarcts in children.

Case report: Two cases of cerebral ischemic infarct due to moyamoya disease in girls aged 7 and 12 are presented. The final diagnosis was established after MR exams and digital subtraction angiography (DSA) of the cerebral arteries.

Conclusions: In spite of rare incidence, the moyamoya disease should be considered as the potential cause of cerebral ischemic infarctions in children. Despite the main role of the DSA in establishment of the final diagnosis, noninvasive neuroradiological examinations become more and more important in diagnostic schedule of moyamoya disease. MR and MR angiography visualize characteristic radiological symptoms, enabling preliminary diagnosis and are the method of choice in control examinations.

Key words: moyamoya • ischemic stroke • imaging

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Background

Progressive idiopathic arteriopathy in childhood called the *moyamoya* disease (m.m.d.) was first described in Japan in 1955 as it occurs most frequently in that region [1]. It rarely occurs in Caucasians [2]. What causes the clinical symptoms observed in course of m.m.d. is the increasing occlusion of the internal carotid artery in supraclinoid segment and its ramifications, i.a. anterior and central brain artery.

Network of collateral circulation, formed compensatively, particularly well developed within the basal ganglia, is called *moyamoya* in the Japanese language [3, 4, 5]. Dominating symptoms include: transient ischemic incidents, less frequently – ischemic and hemorrhagic strokes with paresis, often migrainous headaches [2, 3, 6, 7]. The moyamoya syndrome affects children (12–50%) aged 3–4, as well as 35–40-year-old adults [1, 8]. The pathogenesis remains unclear; several factors, including inflammatory, autoimmunologic and genetic, are taken into account [2, 4, 9, 10, 11, 12]. Although MR and CT with or without vascular program are important for diagnostics, the catheter angiography of carotid arteries remains crucial [3, 8, 13].

Conservative treatment is based on anticoagulants, calcium-channel blockers, vasodilators, corticosteroids, fibrinolitics. Nowadays it is believed that the only effective method of treatment is the operative procedure. Surgical

Figure 1. Girl aged 7: Head CT: ischemic lesions in parietal and occipital regions, edge enhancement after i.v. contrast administration.

methods include: frontal or parietal superficial temporal artery to middle cerebral artery (STA-MCA) bypass surgery, encephalo-duro-arterio-synangiosis (EDAS), encephalo-myo-synangiosis (EMS) [4, 7, 8].

Case report

1st patient: a 16-year-old girl with complicated pregnancy/delivery medical history (premature infant with birth weight of 1700 g) but growing properly. At the age of 10 she started to complain about periodic undiagnosed headaches in frontal region, which were treated tentatively with non-steroid anti-inflammatory drugs with good effects. When the patient was 12 an increasing 5-day hemiparesis occurred on the left side. The MR showed atrophic and angiogenic regions in the left frontal lobe and demyelinating ischemic lesions in white matter of both cerebral hemispheres. Multiple small vessels of tortuous shape were found within the left-side thalamus. In the angioMR decreased circulation in internal carotid arteries was stated, especially in the distal part of left ICA, anterior and middle cerebral arteries. Small tortuous vessels were visualized bilaterally in the middle part. Digital subtraction angiography showed bilateral narrowing of the distal parts of carotid arteries but did not show the right middle cerebral artery. The *moyamoya*-type vascular network of the collateral circulation was observed.

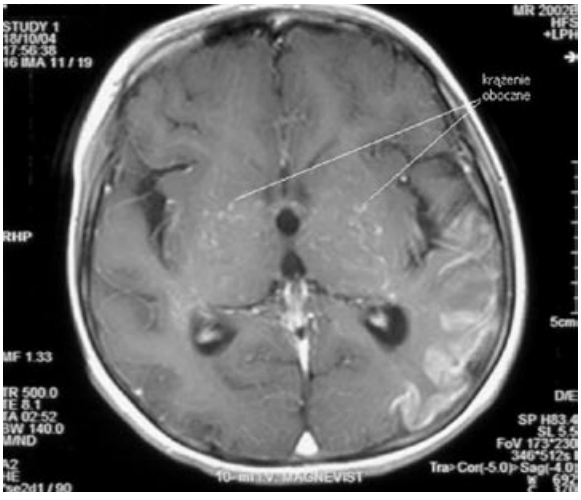

Figure 3. Girl aged 7: Head MR, SE/T1 after contrast administration: evolution of ischemic lesions in left occipital region, collateral vessels visible in basal ganglia.

Figure 2. Girl aged 7: Head MR after 4 weeks, FLAIR image: ischemic lesions in parieto-occipital regions.

After the neurosurgical consultation the patient was qualified for encephalo-duro-arterio-synangiosis, but the parents did not agree for the operation. After 14 days of conservative therapy and intensive rehabilitation the patient was discharged from hospital in good condition with trace hemiparesis. The patient remained under constant neurological ambulatory observation – her state was stable and no more hemorrhagic incidents were observed. At the age of 16 the patient underwent a control angioMR examination of the brain which showed reduction of blood-flow signal in cavernous segments of internal carotid arteries, mainly on the left side, with fragmentarily visible circulation in anterior and middle arteries and multiple small vessels of collateral circulation.

2nd patient: a 7-year-old girl, child of young healthy unrelated parents, with C2, maintained, P1 (C1 – spontaneous abortion in the 2nd month), born on time in spontaneous labor with birth weight of 3200 and 10 points in Apgar scale. Psychokinetic development was normal, milestones were achieved on time. At the age of 7 periodical gait disorders appeared in form of right leg limping, after 6 months the right-side hemiparesis occurred preceded with 2 days of severe headaches. The CT of head showed regions of reduced density within parietal and occipital areas including the white matter and cortex, with marginal enhancement after contrast agent administration (fig. 1). Additionally, the MR visualized signs of decrease in volume of affected areas on the right side and cortical edema in the left parietal lobe.

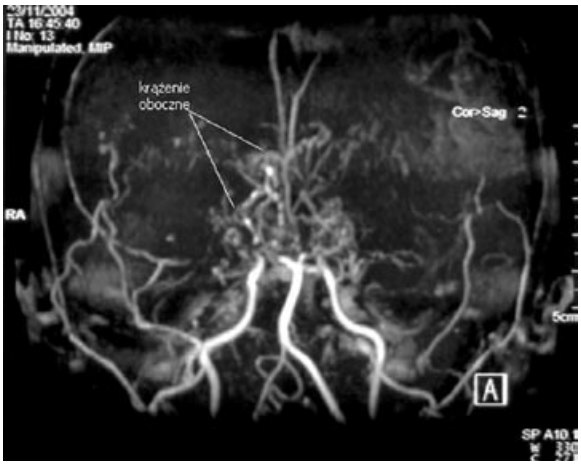

Figure 4. Cerebral arteries angioMR after 8 weeks, a/p projection: lack of flow in medial and anterior cerebral arteries, collateral circulation in the basal ganglia region.

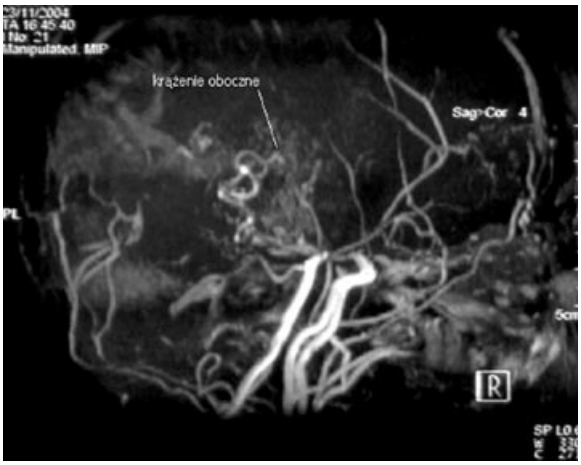

Figure 5. Girl aged 7: Cerebral arteries angioMR after 8 weeks, lateral projection: lack of flow in medial and anterior cerebral arteries, collateral circulation in the basal ganglia region.

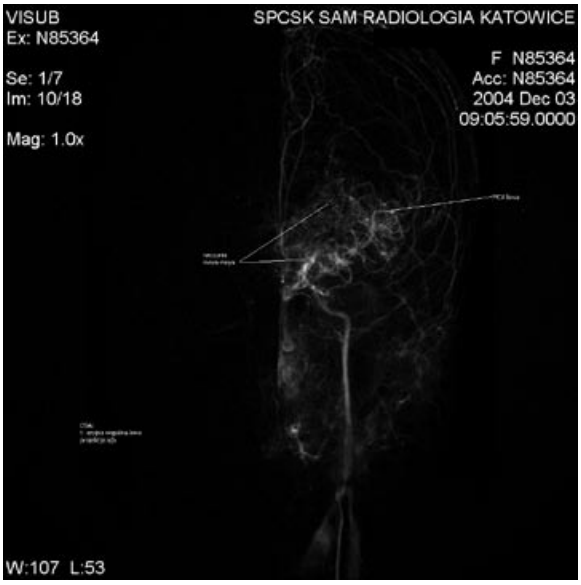

Figure 6. Girl aged 7, DSA of the left CCA, a.p. projection: occlusion of the distal segment of the left ICA, collateral flow to MCA.

Figure 7. Girl aged 7: DSA of the right CCA, a/p projection: occlusion of the distal segment of the right ICA, moyamoya vessels in the basal ganglia.

Within the next 2 weeks patient's condition deteriorated once again – difficulties in swallowing and aphasia appeared. Subsequent MR examinations of head performed after 4 weeks showed evolution of the radiological image of ischemic lesions in parietal regions and the white matter (fig. 2, 3). In angioMR after 8 weeks, the *moyamoya* symptoms were stated in form of small vessels observed in projections of subcortical nuclei and thalamus (fig. 4, 5). Digital subtraction angiography showed occlusion along the internal carotid arteries, lack of flow in the left internal carotid artery above the origin of ophthalmic artery and collateral circulation through those arteries (fig. 6, 7). The middle and anterior arteries were occluded, practically invisible. Multiple focuses of capillary vessels which corresponded with collateral circulation starting from the branches of the external carotid artery, were observed [8, 9]. In the angiography of the left vertebral artery the posterior cerebral arteries were visible only in the P1 segments, and in the later phase small collateral circulation vessels were seen (fig. 10). In EEG generalized paroxysmal lesions were stated.

Conservative treatment was applied along with intensive rehabilitation and logopedic therapy, what resulted in the improvement in child's condition – lower degree of paresis, stronger spontaneous activity, decrease in level of aphasia and regression of attacks. The patient remains under constant neurological care and is systematically rehabilitated.

11 months since the first symptoms had appeared minor motor aphasia, paralysis of the 7th nerve on the left side and trace pyramidal paresis of the left upper limb, still remain. Control MR and angioMR showed no progression of changes.

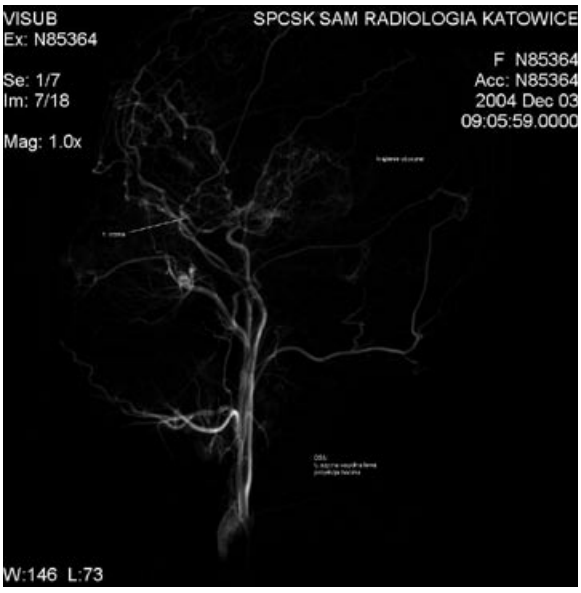

Figure 8. Girl aged 7, DSA of the left CCA, lateral projection: occlusion of the distal segment of the left ICA, collateral flow through the ophthalmic artery.

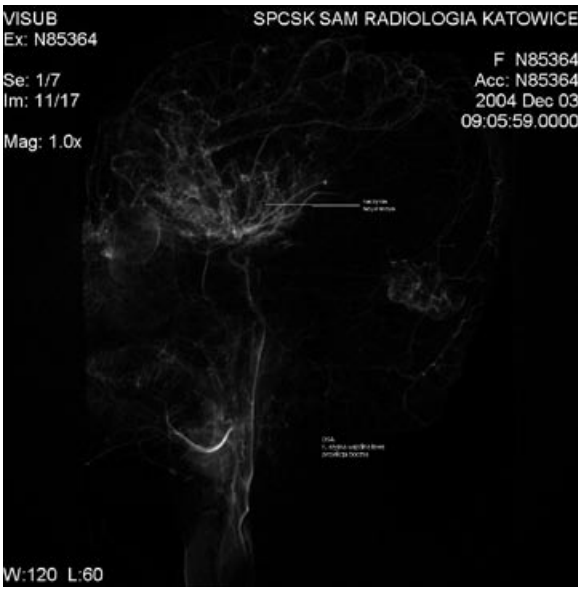

Figure 9. Girl aged 7, DSA of the left CCA, lateral projection: collateral vessels visible in late arterial phase.

In both of the girls we excluded neuroborreliosis, leptospirosis, tick-borne encephalitis, herpes virus infection and tuberculosis on the basis of additional examinations. The results for cerebrospinal fluid were normal, its culture was sterile and the blood-cerebrospinal fluid barrier was leak proof. Additional examinations showed accelerated erythrocyte sedimentation reaction and positive result of Combi test what suggested an autoimmunological disease. Capillaroscopy revealed signs of angiopathy

Discussion

The *moyamoya* recognition is based on the results of imaging examinations. MR is the exam of choice, especially in children, as it visualizes the regions of no signal due to circulation in collateral circulation vessels in basal ganglia. After the contrast agent administration these regions are amplified [14]. MR also reveals past ischemic lesions and the use of EPI DWI sequence enables recognition of early ischemic lesions and differentiation of past and acute lesions. Se/T1 sequences after contrast agent administration are best for visualization of leptomeninx enhancement resulting from the development of collateral circulation net on its surface – the so-called "ivy-sign" [5]. AngioMR with TOF method non-invasively visualizes the occlusion of distal parts of ICA and primary segments of arterial circle, but overvalue of the level of stenosis is possible in case of small vessels, massive occlusion or turbulent flow [14, 15]. That is why it can be difficult to visualize small vessels of collateral circulation in angioMR, especially in adult patients as those vessels atrophy on an advanced stage of the disease [14]. Greater accuracy in recognition of lesions typical for m.m.d. have been reported for the angioMR with contrast enhancement [11, 16].

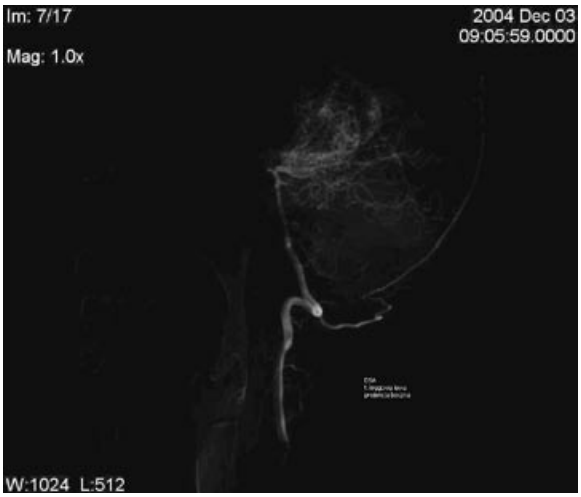

Figure 10. Girl aged 7, DSA of the left vertebral artery: collateral vessels in posterior circulation.

As the arterial morphologic lesions themselves do not enable prognosis of m.m.d., the attention has lately been drawn to the possibilities for assessment of cerebral perfusion in planning the surgical treatment. Study of MR perfusion revealed no statistical dependence between the level of occlusion of the carotid arteries and the risk of stroke, while the reduce and retardation of cerebral perfusion was observed in case of narrowing of the posterior cerebral arteries which get worse with the development of stenosis and lead to perfusion atrophy in PCA occlusion. It is related to a growing risk of stroke. The assessment of cerebral perfusion is also significant for evaluation of surgical m.m.d. treatment results. In view of its non-invasiveness, the MR perfusion is bound to have a wider application in children than the CT, SPECT, PET perfusion, although one ought to be aware of the technical difficulties in its performance and interpretation in *moyamoya* patients [17, 18].

Still, the definite diagnosis of m.m.d. is often based on the result of catheter angiography [15, 17]. In the diagnostic schedule of moyamoya performance of the catheter angiography is advisable after the recognition of *moyamoya* symptoms in MR and

angioMR for a more precise assessment of collateral circulation pathways and determination of the advancement of the disease.

Conclusions

In spite of rare incidence in our country, the moyamoya disease ought to be taken into consideration as a potentail reason of ischemic strokes in children.

MR and angioMR show typical radiological lesions which enable a preliminary diagnosis and are the methods of choice as control examinations; however, precise assessment of cerebral circulation requires the DSA.

Differentiation of the syndrome and *moyamoya* disease requires consideration of clinical image and the results of additional examinations.

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